

Neurologically Handicapped Children

The Role of the Pediatrician in Rehabilitation

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IN THE PAST DECADE much has been learned about the techniques of rehabilitation. The philosophy of rehabilitation, embodying as it does attention not only to the physical but also to the emotional, the social and the educational aspects of the crippling disorders of childhood, has been epitomized in several excellent reviews which have appeared in the pediatric literature within the past decade.^{5,10,12,24} Much emphasis has been given to the "multidisciplinary approach"²⁷ on the basis that the assault on the multiple problems of the severely disabled child—for example, the child having a neurologic handicap—requires a battery of skills too diverse to be encompassed by any individual physician.

In recent years numerous elaborate rehabilitation centers have come into being, staffed by large numbers of medical specialists with coworkers from such other professions as psychology, social work, speech pathology and audiology, physical therapy and occupational therapy. These are generally located in the more densely populated urban areas. In addition, educators have given much attention to the problems of the handicapped child, particularly to those of the child having a neurologic impairment, with the result that in many areas such children are segregated in special schools for the disabled throughout their scholastic experience.

In the consideration of problems of the child having a neurologic impairment, much emphasis has been given to methods of evaluation and management. Techniques for early case-finding,²⁶ for classification of the neurologic deficit,² for detection of associated damage to the visual and acoustic apparatus,^{7,22} for assessment of disability in the areas of language and communication^{1,13} and for the psychologic appraisal of the brain-damaged child^{14,18} are given extended discussion. In the fields of physical therapy⁶ and of occupational therapy,¹⁷ those concerned with the practical aspects of assisting the child toward the development of specific neuromuscular skills have evolved elaborate regimens which can be employed only by those having a highly specialized background of training. Educators have

• In the application of the broad services now available to assist a child having a major neurologic impairment, the pediatrician occupies an important role owing to his ability to consider the problem of the handicapped child in the context of his specialized knowledge of the developmental process. He thus has a large responsibility for interpretation of the problem to the child, to the parents and to his professional colleagues and for guidance of the rehabilitation regimen within the limits of the child's developmental readiness for new experiences.

The pediatrician has the opportunity to contribute significant clinical observations which may provide stimuli for future basic research and to exercise his skill as a practitioner of preventive medicine.

Goals for the future achievement of the child having a major neurologic impairment must be set realistically and with great caution.

published extensively, often in conjunction with psychologists, on the particular learning disabilities encountered in children having impairments of the central nervous system and on the special techniques of education that have proved most rewarding in this field.^{9,23}

Methods for parent counseling, and in particular group therapy techniques,^{16,19} have been tested with parents of children having a variety of handicaps and have proved fruitful in assisting parents toward the development of insight into the dynamics of their relationships with their handicapped and frequently emotionally disturbed children. In addition, the sociologists and the cultural anthropologists²⁴ have asked that due consideration be given to community and cultural factors as they relate to the individual undergoing what now appears to be the almost overwhelming experience of becoming rehabilitated.

In view of the special knowledge required, it would be small wonder if the pediatrician, looking upon himself now as a generalist, should feel abashed and even at times bewildered. When considering the complexities that have been devised for the study and care of patients handicapped by neurologic deficits, one can visualize the pediatrician asking himself in the dead of night, "Am I a sociologist, a social worker, a psychologist? Am I a neurologist, a physiatrist? Where do I fit in? What is

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my role?" He may well reflect that the "multidisciplinary approach," although it has proved useful in bringing to light many specialized aspects of the complex problem of central nervous system impairment, has also built within it the potential for creating in his patients and their families an even larger degree of confusion than he had felt momentarily when he paused for self-scrutiny. The pediatrician must remind himself that only he, of all the professional people with whom his patient may have contact, can view the child's total situation with the perspective and wisdom that a specialized knowledge of the process of development permits. With his knowledge of the ramifications of the developmental process the pediatrician is in a position to evaluate the dynamic significance of one aspect or another of defective neurologic function. He is the only one in a position to interpret to the parent the significance of the neurologic impairment as it relates to aptitude of intellectual function, perceptual ability and learning capability and how these in turn relate to emotional maturity and to the image that the child has formed of himself. It is his large responsibility to interpret these matters to the parent in the light of his own knowledge of the complexities of the developmental process.

Although the pediatrician here finds himself in the role of a teacher to the parent, he has another more primary role, even more important. With his knowledge of the dynamic process of growth and development of the central nervous system, he is in the unique position of capability for assigning values to the various manifestations of neurologic dysfunction which may be manifested by reflex activity, by lack of cortical inhibition, by ataxia, dystonia or the like. His sophistication in matters pertaining to the

developmental process permits him to relate laboratory and electroencephalographic findings to the physical and psychologic attributes of his patient as he views him in a given moment of time. This knowledge of the basic processes underlying the total development of the child and, more specifically, what is known of the biochemical development of the central nervous system, permits the pediatrician to recognize his patient's readiness or lack of readiness for new experiences and specifically for any new mode of therapy, training or scholastic instruction that may be planned. Thus, recognizing the importance of tailoring treatment regimens and scholastic programs in such a fashion that they will be the most appropriate for his patient's developmental level and for his emotional readiness for new experiences, the pediatrician will be in constant touch with the physical therapist, the speech therapist and the teacher, truly coordinating their activities in a fashion that will permit his patient to make the maximum gains from the specialized help that is offered him. Likewise, his knowledge of the intimate parent-child interaction will permit him an understanding of the forces pertaining in the home which will permit optimal developmental progress on the one hand, or, in a situation in which a disturbed relationship exists, an interference with the developmental process and consequent complications in the therapeutic or learning situation.

CAUSES OF NEUROLOGIC IMPAIRMENT

The major disabilities of the central nervous system seen in infancy and childhood may be divided into those having their origin in a genetic abnormality of the germ plasm or otherwise originating

TABLE 1.—*Classification of Causes of Neurologic Impairment in the Developing Organism*

Period of Onset	Underlying Disturbance	Examples
Prenatal	1. Genetic disorder	a. Gargoylism b. Lipoid storage diseases c. Galactosemia d. Phenylketonuria
	2. Congenital malformation	a. Myelodysplasia b. Craniosynostosis
	3. Infection	a. Toxoplasmosis b. Maternal rubella
Perinatal	1. Infection	a. Sepsis
	2. Trauma, hypoxia or vascular injury	a. Spastic quadriplegia or hemiplegia
	3. Endogenous neurotoxic agent	a. Hyperbilirubinemia
Postnatal	1. Metabolic	a. Porphyria
	2. Nutritional	a. Convulsive state related to pyridoxin deficiency
	3. Infection	a. Viral encephalitis b. Suppurative meningitis
	4. Trauma, hypoxia or vascular injury	a. Hemiplegia, quadriplegia
	5. Neoplasm	a. Glioma, ependymoma
	6. Exogenous toxic substances	a. Lead encephalopathy b. Drug toxicities

in prenatal life, those which originate during the perinatal period and those having their beginnings at some point in the postnatal period. These may, in turn, be divided into those expressing themselves in a metabolic dysfunction (of which a majority have an essentially genetic basis); those manifesting themselves by congenital malformation (of which some have a clearly genetic basis and some appear to be related to environmental insult during intra-uterine life); those related clearly to infection; those related to trauma or vascular damage; those related to specific nutritional lack; those related to endocrine abnormality; those due to neoplasms of the brain; those due to endogenous neurotoxic substances; and those in which central nervous system dysfunction is due to the action of an exogenous poison. There is considerable overlap in these categories. Table 1 provides a far from complete listing of examples of these several categories of disorders of the central nervous system.

The pediatrician has the opportunity to play the most significant role in the detection of brain injury. Through his close relationship with child and family he is apt to be the first to become aware of a deviation from the smooth course of normal development, and so to arrive at a diagnosis of impairment in neurologic function. Beyond this, the pediatrician has an even more exciting opportunity. With him lies the potential, through his critical clinical observations, for uncovering previously unrecognized basic processes which may lead to central nervous system dysfunction. An example will suffice: In 1954 Hunt and his collaborators¹¹ observed a young infant having intractable convulsive seizures. Through an elegant plan of clinical study they were able to define that this baby's convulsive difficulty was based on what they characterized as "pyridoxine dependence." The administration of pyridoxine controlled the seizures. Later others,³ observing convulsive phenomena in infants, fed a proprietary milk mixture which lacked pyridoxine, confirmed Hunt's earlier observation and were able to define the requirement of the infant for this essential nutrient substance. In the meantime the biochemists,^{21,25} studying the metabolic effects on the brain of pyridoxine deprivation, were able to define that the occurrence of epileptiform seizures in the experimental animal so deprived is correlated with a specific decrease of glutamic decarboxylase activity (for which pyridoxine serves as a coenzyme) with consequent failure in the formation of γ -aminobutyric acid, an amino acid found only in the tissue of the central nervous system. Further basic research has disclosed that in all cases of epileptiform seizures studied, both in the experimental animal and in brain excised from human beings having focal epilepsy, there is evidence for involve-

ment of glutamic acid itself or of γ -aminobutyric acid.

A further opportunity of the pediatrician has to do with his role in early case-finding. Here, through the accurate identification of a metabolic disorder in the young infant, at a stage prior to the development of major significant brain damage, he may be able to control the metabolic dysfunction through dietary or other means and so permit the child unimpaired brain development which would otherwise have been denied. The employment of a lactose-free diet in galactosemia¹⁵ and of a dietary low in phenylalanine in phenylketonuric patients⁴ are examples familiar to everyone. These examples point up the growing importance of improvement of our understanding of genetic disturbances.²⁰ A tool which would permit the identification of the heterozygote in either of the situations cited here, or in many of the other traits transmitted by an autosomal recessive gene, would be invaluable in permitting the early identification of the homozygous individual and the application of appropriate measures for management where these are feasible.

GOALS IN THE REHABILITATION

In planning a program for a child having a major neurologic deficit, multiple areas must be considered, as has been pointed out earlier. These include the management of the physical aspects of the disability with appropriate consideration being given to the child's intellectual capacity, to his level of emotional maturity and perhaps particularly to the image he has of himself, as well as to the fashion in which he relates to his peers and to adults. His educability, and particularly the fashion in which he perceives spatial relationships, as well as his manner of performance in certain disciplined activities such as reading, writing and the use of numbers, requires careful consideration. Lastly, detailed knowledge of his family and of his home situation are essential. The impact upon the parents and indeed upon the entire family, as it can be defined in terms of economic stress, of guilt and of anxiety, of the ambivalence that the parents may feel toward the handicapped child in terms of overprotection on the one hand and rejection on the other, is a matter which cannot be underestimated. Here the collaborative services of a skilled social worker are invaluable to the busy physician. Not only can the social worker be useful in interpreting to the family the interplay of feelings which predictably occur in this sort of situation, but her knowledge of community resources available to the physician, which will assist him in meeting the needs of the family, can be most helpful.

TABLE 2.—Intellectual Competence of Brain-Damaged Patients^a

Intellectual Competence Category	Pyramidal Involvement Only		Extra-pyramidal Involvement Only	
	No.	Per Cent	No.	Per Cent
Superior.....	12	4	11	12
Average.....	53	19	31	33
Borderline.....	71	26	20	22
Defective.....	62	23	19	20
Low-grade defective.....	75	28	12	13
Total.....	273	100	93	100

Perhaps the most serious pitfall that is met in the process of rehabilitation is that of setting unrealistic goals. This is well illustrated in Table 2, which is an adaptation of Bronson Crothers's⁸ data on a large group of brain-damaged children who were observed carefully, with repeated determinations of intellectual functioning, psychologic aptitude and performance over an extended period.

The data speak for themselves. They are, to say the least, depressing. It seems obvious that, in this group of children where almost half are patently mentally defective, the goals to be anticipated in terms of future achievement must be set with great caution. The parents must be carefully and gradually prepared to view the future with realism. The rate of salvage, placed in terms of the likelihood of future independence and important contribution to society, will be low. A similarly cautious prognosis must be given with regard to the child who has developed manifestations of severe personality disturbance with relation to his disability.

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